

RHEUMATIC FEVER

Disease name

Disease	Indicate whether this is an initial (i.e. first) episode of rheumatic fever or a recurrent episode (an episode in a person with known past history of acute rheumatic fever (ARF) or previously diagnosed rheumatic heart disease).
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Basis of diagnosis

Jones criteria Jones criteria			
The Jones criteria for the diagnosis of ARF divide the clinical features into major and minor manifestations, based on their prevalence and specificity. The diagnosis of rheumatic fever requires evidence of a preceding group A streptococcal infection, and the case classification is then based on the presence of major and minor manifestations. <table> <tr> <td> Major manifestations: - carditis - polyarthritis or aseptic monoarthritis - chorea - erythema marginatum - subcutaneous nodules </td><td> Minor manifestations: - fever $\geq 38^{\circ}\text{C}$ - raised acute phase reactants: erythrocyte sedimentation rate (ESR) ≥ 50 mm/hr or C reactive protein (CRP) ≥ 30 mg/L - polyarthralgia - prolonged PR interval </td></tr> </table> Notes: - if carditis is present as a major manifestation, a prolonged PR interval cannot be considered an additional minor manifestation; - if polyarthritis or monoarthritis is present as a major manifestation, polyarthralgia cannot be considered an additional minor manifestation; - chorea can be a stand-alone manifestation for ARF diagnosis provided other causes are excluded.		Major manifestations: - carditis - polyarthritis or aseptic monoarthritis - chorea - erythema marginatum - subcutaneous nodules	Minor manifestations: - fever $\geq 38^{\circ}\text{C}$ - raised acute phase reactants: erythrocyte sedimentation rate (ESR) ≥ 50 mm/hr or C reactive protein (CRP) ≥ 30 mg/L - polyarthralgia - prolonged PR interval
Major manifestations: - carditis - polyarthritis or aseptic monoarthritis - chorea - erythema marginatum - subcutaneous nodules	Minor manifestations: - fever $\geq 38^{\circ}\text{C}$ - raised acute phase reactants: erythrocyte sedimentation rate (ESR) ≥ 50 mm/hr or C reactive protein (CRP) ≥ 30 mg/L - polyarthralgia - prolonged PR interval		
Major manifestations Major manifestations	Ideally, obtain information on all of the clinical features listed. If the feature was present, record by ticking the “Yes” box. If not, tick the “No” box. If not known or unavailable then tick the “Unknown” box.		
Minor manifestations Minor manifestations	Ideally, obtain information on all of the features listed. If the feature was present, record by ticking the “Yes” box. If not, tick the “No” box. If not known or unavailable then tick the “Unknown” box.		

Supporting laboratory criteria for streptococcal infection	
Evidence of preceding group A streptococcal infection	Indicate whether there is evidence of a preceding group A streptococcal (GAS) infection. If not known or unavailable then tick the “Unknown” box. If “Yes”, provide details for each laboratory test. If the laboratory result provides evidence of a preceding GAS infection tick the “Yes” option, if not, tick the “No” option. If the test was not carried out tick “Not Done”. If not known or unavailable then tick the “Unknown” option. The current antibody titre cut-off levels are listed in the case classification section (laboratory criteria).
Antibody titre results	If serological antibody tests were done, specify the antibody titre results for the first and second (if applicable) tests. The titre results should be entered regardless of whether they meet the current level of evidence for preceding streptococcal infection. Specify the date of each test.

Previous history of rheumatic fever (recurrences only)	
Previous episodes	Indicate the number of previously diagnosed episodes. Note the dates when the first and most recent previous episodes occurred and specify the hospitals where the patient was diagnosed. If either of the dates is unknown, then tick the “Date unknown” box.
Evidence of previous rheumatic heart disease	Indicate whether there was evidence of previous rheumatic heart disease prior to this episode. If there was evidence, tick the “Yes” box, if not, tick the “No” box. If not known or unavailable tick the “Unknown” box.

Classification

Classification	Under investigation - a case that has been notified but information is not yet available to classify it as suspect, probable or confirmed. Suspect - <u>Initial episode</u>; requires all of the following: • strong clinical suspicion of ARF but insufficient signs and symptoms to fulfil diagnosis of confirmed or probable ARF • no other alternative explanation for the clinical presentation. <u>Recurrent episode</u>; requires all of the following: • strong clinical suspicion of ARF but insufficient signs and symptoms to fulfil diagnosis of confirmed or probable ARF • no other alternative explanation for the clinical presentation • a prior confirmed episode of ARF or rheumatic heart disease.
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Probable -

Initial episode; requires **all** of the following:

- one major and one minor manifestation
- evidence of preceding group A streptococcal infection (see laboratory criteria).

Recurrent episode; requires **all** of the following:

- one major and one minor manifestation
- evidence of preceding group A streptococcal infection (see laboratory criteria)
- a prior confirmed episode of ARF or rheumatic heart disease.

Confirmed -

Initial episode; requires **all** of the following:

- two major manifestations OR one major and two minor manifestations
- evidence of a preceding group A streptococcal infection (see laboratory criteria).

Recurrent episode; requires **all** of the following:

- two major manifestations, OR one major and two minor manifestations, OR three minor manifestations
- evidence of a preceding group A streptococcal infection (see laboratory criteria)
- a prior confirmed episode of ARF or rheumatic heart disease.

Not a case – a case that has been investigated and subsequently found not to meet the case definition.

Laboratory criteria for classification

For ARF cases that have carditis as a major manifestation:

- GAS serology – rising titres OR a single antibody level above the upper limits of normal (ASO $\geq$ 444 or ADB $\geq$ 400 IU/ml)
- positive GAS throat swab culture in the last 4 weeks
- positive GAS rapid molecular test result on a throat swab in the last 4 weeks

For ARF cases that do not have carditis as a major manifestation:

Confirmed (e.g. meets the major and minor criteria)-

- GAS serology – rising titres OR a single antibody level above the upper limits of normal (ASO $\geq$ 444 or ADB $\geq$ 400 IU/ml)

Probable (one major and one minor manifestation)-

- GAS serology – rising titres OR a single antibody level above the upper limits of normal (ASO $\geq$ 444 or ADB $\geq$ 400 IU/ml)
- positive GAS throat swab culture in the last 4 weeks
- positive GAS rapid molecular test result on a throat swab in the last 4 weeks

Risk factors

Recent sore throat	
Sore throat history	Indicate whether there was a history of sore throat in the 4 weeks prior to the hospital admission or the clinic visit where the rheumatic fever diagnosis was made. If yes, indicate whether the case sought medical attention for their sore throat and provide details of the antibiotics prescribed. If not known or unavailable then tick the “Unknown” box.
Family history of rheumatic fever	
Family history	Indicate whether any family members have had rheumatic fever in the past. If yes, state their relationship to the case (e.g. mother, sister etc.). If not known or unavailable then tick the “Unknown” box.

Management

Case management	
Case on rheumatic fever register (initial episodes)	For initial episodes, indicate whether the case was placed on a rheumatic fever register or secondary prevention care coordination system. If not known or unavailable then tick the “Unknown” box.
Case already on rheumatic fever register (recurrent episodes)	For recurrent episodes, indicate whether the case was already on a rheumatic fever register or secondary prevention patient management system. If “Yes”, state the name of the register or PMS (i.e. what district or region and organisation). If not known or unavailable then tick the “Unknown” box.